Modern Concepts of Cardiovascular Disease

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CALCIFIC AORTIC STENOSIS*

Calcific stenosis of the aortic valve is a subject of much theoretical and practical interest, even though we do not understand its pathogenesis and do not yet have a really satisfactory method for removing the mechanical impediment which threatens ultimately to throttle the circulation and snuff out the patient's life. With surgical research on the move, it is likely that newer methods will improve operative results and reduce mortality. Even so, the mechanical ruin of severely stenotic valves limits the degree of restoration towards normal which can ever be attained. Until we are able to prevent the occurrence of aortic stenosis, the hope for the future may lie with those who can devise mechanical contrivances to replace the valve, rather than along the lines of the repair work of the mining engineer. But even when we can give no help by surgical means, the diagnosis should be made so that appropriate medical therapy can be used and the prognosis understood.

The history of aortic stenosis as a medical problem clearly reflects the general state of medical knowledge over the last hundred years. The diagnosis became possible not much more than a hundred years ago, as the normal heart sounds were slowly separated from abnormal sounds or murmurs. By the end of the last century, aortic stenosis was recognized on the basis of a systolic murmur heard best at the base of the heart, usually accompanied by a thrill. The murmur might be transmitted, especially upward into the neck. Only when routine recordings of arterial blood pressure became established in practice was emphasis put on the *pulsus tardus et parvus*, in spite of a very ancient pulse lore. A slowly rising little pulse wave, readily perceived in some instances of aortic stenosis by the well-trained finger tips, and now recorded by the intra-arterial needle, is not the usual finding.[1] This classic diagnostic requirement of small pulse and reduced pulse pressure has been a distraction rather than a help, since it is the exception, not the rule.

In the period before and after World War I, the systolic murmur fell into disrepute. Physiologic methods opened new vistas of evaluating function, and the electrocardiograph began to elaborate on the facts learned from the polygraph and thus disclose the secrets of the arrhythmias. Interest in physical signs waned. The popular and increasingly prevalent problems of coronary artery disease, at times devoid of any physical sign at all, caught the fancy of physicians. A pang of angina or a crisis of coronary thrombosis might be devoid of any clue from physical diagnosis. Interest in aortic stenosis, which had never become extinct but merely overshadowed, began to revive with the emergence of the great school of clinical cardiologists in this country. Cabot, Christian, White, Levine and their colleagues and disciples brought back for clinical emphasis syncope, heart pain, sudden death, restored interest in the basal systolic murmur which is transmitted upwards, the thrill, large heart and slow pulse. The disorder was so well described that the clinical picture again became static.

The history of medicine has many examples of repeated cycles of progress, halt, and regression in our understanding of particular medical problems. Here is what happens: The classic or textbook picture is merely the average of a writer's experience, blended with what he has read. If the description is convincing, or the author is an "authority," it freezes into dogma. Then series of cases drift into "the literature". Selected clinically on the basis of the diagnostic requirements of the classic picture or its close affiliates, the findings are added up and turn out to verify the prerequisites. Cases which have been diagnosed as aortic stenosis because of a basal systolic murmur, thrill and small pulse, do, in fact, have them. One has defined the diagnostic criteria. Then it is dogmatized that aortic stenosis is characterized by signs found in patients selected on that basis. Each paper consolidates the position and increases the clinician's illusion of understanding. A step towards correction comes from discovery of false positives and false negatives. In such a disease as aortic stenosis this can come only when the autopsy discloses the unsuspected lesion, or when the lesion, strongly suspected or diagnosed with assurance, is not found at autopsy.

Some years ago, after we had missed the

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diagnosis of aortic stenosis twice in one week, Kumpe and I decided to see what might be wrong with us or our ideas of the disease.[1] We reviewed 107 autopsied cases of aortic stenosis which were not complicated by mechanical fault of other valves. We were aware that a retrospective study from autopsy records has bias. How representative is the hospital population of the community at large; what percentage of patients die; of these what proportion are autopsied and what bias does current interest of the autopsy seeker introduce? Such valid objections are counterbalanced in part by the fact that, where the clinical diagnosis is unsuspected or wrong, the actual findings will not have been warped to fit a required diagnostic pattern.

Our findings came as a surprise to us. They showed how artificial had become the criteria for suspecting the presence of aortic stenosis. Our major findings and conclusions have been verified and extended by subsequent studies.[2,3] Some of the salient features of these and other reports may be summarized as follows:

Age—Calcific aortic stenosis is a disease of the elderly, though the lesion may occur in children. In the 70's it is 10 to 15 times more prevalent, in the 30's less than one-fifth as prevalent as it would be if the risk were the same at all ages. The implication is clear in a nation of aging citizens.

Sex—Men outnumber women nearly 3 to 1.

Rheumatic Fever—The importance of rheumatic fever may be estimated by noting that in our studies in Cincinnati and in Iowa a positive history was usually obtained in about half the patients with aortic stenosis while a positive history of rheumatic fever was usually found in about the same proportion in younger persons with mitral stenosis. Thus rheumatic fever seems to be the major precursor. Karsner and Koletsky[4] believed that 98 per cent of their 200 autopsied patients had had rheumatic fever. Operations on the mitral valves, giving us an opportunity to get biopsies of the heart, suggest that the rheumatic process may go smouldering on without clinical signs. Such continuous activity might help explain the aortic valve calcifications which may not occur until the patients are in their 60's, 70's, or 80's. No doubt some lesions arise from various kinds of arteriosclerosis and others as a consequence of the deformity of a congenitally bicuspid valve.

Autopsy Findings

Calcific aortic stenosis may vary from a relatively mild lesion with calcification, partial obstruction of the valve aperture and some mobility of the valve, to a "fish-mouth" orifice with a funnel-shaped valve deformity pointing upward, frozen as solid as stone and on rare occasions containing actual bone. Sometimes lines of cleavage can be made out; in others they are lost in the distorted frozen mass. If the plane of cleavage can be found, sometimes at operation the valves can be opened along lines of adhesion and a partially effective hinged trap door arrangement may result. No blind approach can ever be very reliable, particularly when an instrument resembling an umbrella is inserted into the stenotic aperture from below and then opened. At autopsy, in approximately one-half the instances, fusion of the cusps is found together with nodules and every variety of deformity. Atheromatous ulcers may occur. Surprisingly enough, bacterial endocarditis is not rare.

Heart Weight

Generally, in calcific aortic stenosis, the heart is considerably larger in men than in women. The size of the heart and the thickness of the left ventricle tend to increase with increasing severity of the stenosis. Severe sclerosis of the aorta is common. It tends to be mild in the ascending aorta and increase progressively as one proceeds distally. Coronary artery sclerosis, with or without old infarction or fibrosis, may be found in as many as 25 per cent of the hearts with calcific aortic stenosis. Aside from the heart, the findings are those associated with chronic congestion. In some instances, ischemic changes occur in the central nervous system and are probably aggravated by repeated periods of ischemia and anoxia.

In reviewing all autopsied cases of aortic stenosis found in the records of the Pathology Department of the State Univerity of Iowa, a survey still in its early stages, Jamison and I[5] have found that with increasing interest in aortic stenosis, the percentage of correct diagnosis has risen to well over 50 per cent. The likelihood of the diagnosis being made depended largely on whether the patient was on a hospital service where a careful and complete physical examination and detailed history were recorded.

Symptoms

In aortic stenosis, congestive failure is the commonest cause for consulting a physician or being admitted to a hospital.

Chest pain, sometimes typically anginal and sometimes not related to obvious increased demands on the heart, is common. At least in some patients with aortic stenosis, pain in the chest occurs after exercise, not during it. It is more commonly referred to the right side of the chest or to begin on the right side of the chest than in persons with classic angina. Furthermore the symptom is related more closely to the severity of the stenosis than to the degree of coronary sclerosis as found at autopsy.

Signs of cerebral ischemia, varying from transient flashes of faintness to syncope, are characteristic. Intermittent focal paraiyses, bizarre mental behavior and fits of weakness may occur. A few persons fall and are hospitalized after fractures or head injuries.

Physical Signs

There is no regular pattern of *pulse rate* though tachycardia is common, especially in the presence of congestive failure. *Respiration* is likely to be rapid for the same reason. Contrary to expectation, there is no characteristic arterial blood pressure. One may have systolic or diastolic hypertension or both. The diastolic pressure

may be low or even zero. The so-called classic picture of slightly lowered systolic with normal diastolic pressures, giving a low pulse pressure, is unusual.

Examination of the Heart—Clinical examination usually discloses the heart to be enlarged though it may not be early in the disease. A loud harsh basal systolic murmur heard best over the aortic area is the commonest sign. Transmission of the sound depends more on the loudness of the murmur than on the direction of blood flow or the course taken by the great vessels. Apical systolic murmurs may be louder than basal murmurs. Thus the major murmur may appear to arise in the region of the apex and mitral regurgitation be suspected. A loud aortic diastolic murmur may distract the observer into thinking that aortic regurgitation is the only or major defect. In general thrills, being the tactile percept of which the murmur is the auditory counterpart, are usually felt if there is a loud murmur because of intense vibration. Muffling or complete absence of the "aortic valve closure sound" is a helpful confirmatory sign, but a loud second sound may be heard in the second intercostal space on the right even when the valve can have no mechanical action as a valve. This may be a sound transmitted from the pulmonic valve. Perhaps diastolic impact of blood in the aorta produces it. Irregularities of rhythm are fairly common, but auricular fibrillation is not nearly as common as when there is isolated mitral stenosis or mitral stenosis associated with aortic stenosis. There are no characteristic laboratory clues; although calcification of the aortic valves is found many times by careful fluoroscopic examination, it is rarely observed on routine x-ray films. Often there is electrocardiographic and roentgenographic evidence of left ventricular hypertrophy. The physical signs of congestive failure are common. Congestive failure, chest pain, and syncope may attend the course of the patient while under observation and in the hospital. A syndrome, sometimes mistaken for acute myocardial infarction, may occur in which a crisis of sweating, cyanosis, restlessness, confusion, sometimes flushing and sometimes pallor, may begin abruptly and either lead to an early death or be associated with a slow recovery. Typical or atypical myocardial infarction is not rare as a complication since aortic stenosis confers no immunity to coronary atherosclerosis.

Sudden Death

Sudden death may occur in many ways. Few who write of it have bothered to say what they mean by "sudden." But in careful observations of the process and act of death in people with aortic stenosis, the so-called instant "physiologic" death is not nearly as likely to occur as it is after acute myocardial infarction or in people with severe coronary artery disease and angina.[6] In aortic stenosis it is fairly common for the death scene to continue over a period of a few minutes. If a person dying in this manner is observed intently, ordinarily one finds indications that the heart stops before other functions play out. Respirations are likely to continue for several moments. Consciousness may remain for a short time during which the patient may call out in pain. There may be a short period of constricting field of vision, apprehension and deep respiratory efforts with air hunger. Cyanosis develops and it continues while dissolution proceeds slowly. Anguish, purposeless movement, sometimes focal weakness with tonic and rarely clonic convulsions may be noted. Such a form of dying is not especially common in persons with aortic stenosis who have had a tendency to have syncope attacks. This is in contradistinction to the risk of instant "physiologic" death in persons with acute myocardial infarction but without valve disease. In them abrupt, total and instantly complete death is much commoner in those who have had many faints in the past.[6] The question of sensitivity of the carotid sinus needs to be studied and compared in persons with coronary artery disease alone and those with aortic stenosis alone. Many such points need further detailed observation during life to see how regular are the trends suggested by the retrospective study of what had happened to patients who came to autopsy.

Treatment

The treatment consists of the usual management of heart failure, which, however, is none too satisfactory. Harken has devised a new aortic approach for digital and instrumental fracture of the valves. Brilliant success from finger, or finger guided instrumental cleavage of the frozen valve will be the exception because even when the commissural fusion is broken, the densely calcified valve may remain relatively immobile. The fracture may go beyond the desired degree and even tear the aortic wall. Finally aortic regurgitation may be added to the burden of an already crippled heart. It is the best method available but is not yet satisfactory. By the judicious use of radioactive iodine, thyroid function can be damped down. The patient exchanges some symptoms of aortic stenosis for those of hypothyroidism. Real improvement may occur, and the patient better tolerates the symptoms which persist.

At the present time there is great need of further study of clinical cases to find out: (1) the natural history of the disease in its early and relatively asymptomatic stages; (2) the minimal criteria for diagnosis; (3) the optimal use of measures aimed to combat congestive failure and measures to reduce metabolic requirements such as medical thyroidectomy; (4) the definitive place of the surgical attempt to pry open deformed, stenotic valves and the effort to reconstruct a new valve or insert an artificial one. Only when we rearrange our ideas on the basis of extensive experience from autopsies and then go back and study patients on the ward with new ideas in mind, can we hope to make any definite progress in such a perplexing disorder.

Acknowledgement

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The opinions and conclusions expressed herein are those of the author and do not necessarily represent the official views of the Scientific Council of the American Heart Association.

THE ANNUAL MEETING
and
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of
THE AMERICAN HEART ASSOCIATION
October 27 through November 2, 1956
***Headquarters:* Netherland Plaza Hotel**
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FRIDAY, OCTOBER 26	Advance Registration (3 P.M. to 10 P.M.)	Netherland Plaza

MEETING SCHEDULE

FRIDAY, OCTOBER 26	Special Evening Session Instrumentation, Electrocardiography, Vectorcardiography & Ballistocardiography	Netherland Plaza
SATURDAY, OCTOBER 27	29th Annual Scientific Sessions	Cincinnati Music Hall
through	Second Annual Program of Council on Community Service and Education	Cincinnati Music Hall
MONDAY, OCTOBER 29	Program of the Council on Rheumatic Fever and Congenital Heart Disease	Cincinnati Music Hall
TUESDAY, OCTOBER 30	A. H. A. Assembly Panel Meetings	Netherland Plaza
WEDNESDAY, OCTOBER 31	Annual Meeting of A. H. A. Assembly—elections—panel reports	Netherland Plaza
WEDNESDAY, OCTOBER 31 through FRIDAY, November 2	Annual Meeting of the Staff Conference of Heart Associations	Netherland Plaza

SOCIAL FUNCTIONS

SUNDAY, OCTOBER 28	Community Service & Education Luncheon	Netherland Plaza
	A. H. A. Annual Dinner Dance—presentation of gold heart awards	Netherland Plaza
TUESDAY, OCTOBER 30	A. H. A. Assembly Luncheon	Netherland Plaza
SATURDAY, OCTOBER 27 through TUESDAY, OCTOBER 30	For the Ladies: Luncheons, fashion shows, conducted tours, etc.	Hospitality Booth—Netherland Plaza

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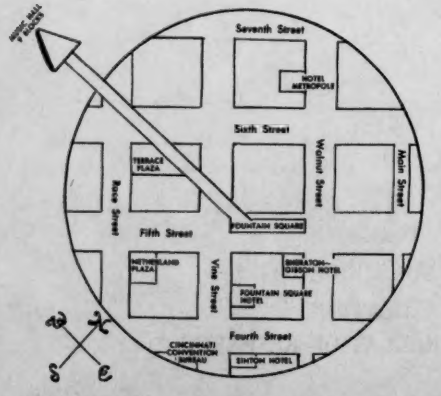

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